

General

Title

Sickle cell disease (SCD): percentage of children younger than 18 years of age identified as having SCD and on a chronic transfusion program who received monitoring of hemoglobin S levels immediately prior to each transfusion during the measurement year.

Source(s)

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). Basic measure information: hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease. Ann Arbor (MI): Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium; 2014 Apr. 39 p.

Measure Domain

Primary Measure Domain

Clinical Quality Measures: Process

Secondary Measure Domain

Does not apply to this measure

Brief Abstract

Description

This measure is used to assess the percentage of children younger than 18 years of age identified as having sickle cell disease (SCD) and on a chronic transfusion program who received monitoring of hemoglobin S levels immediately prior to each transfusion during the measurement year. A higher proportion indicates better performance, as reflected by appropriate treatment.

Rationale

Approximately 2,000 infants are born with sickle cell disease (SCD) in the United States each year, a condition that occurs predominantly in people of African and Hispanic descent. Among children with SCD, approximately 11% experience a stroke by 20 years of age, and another 13% show evidence of silent infarcts (injuries sustained by the brain without clinical symptoms). This damage to the central nervous

system is a devastating complication of SCD. It occurs most frequently in children with the hemoglobin variants associated with sickle cell anemia, the most serious form of SCD. Prevention is a high priority. Regularly scheduled, ongoing blood transfusions have been found to reduce the percentage of sickle hemoglobin (Hb S) to 30% of total hemoglobin concentration. This reduction in Hb S helps prevent a first stroke in high risk patients and recurrent stroke in patients who have already had a first stroke. Chronic transfusion therapy has also been shown to ease other complications related to SCD. Clinical guidelines indicate that chronic transfusion protocols should include regular monitoring of the Hb S level before each transfusion. However, there are no existing quality measures regarding the pre-transfusion monitoring of Hb S in children with sickle cell anemia on a chronic transfusion program.

Evidence for Rationale

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). Basic measure information: hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease. Ann Arbor (MI): Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium; 2014 Apr. 39 p.

Primary Health Components

Sickle cell disease (SCD); sickle cell anemia; hemoglobin S (Hb S); blood transfusion; infants; children; adolescents

Denominator Description

The eligible population for the denominator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program during the measurement year (January 1 to December 31) (see the related "Denominator Inclusions/Exclusions" field).

Numerator Description

The eligible population for the numerator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of hemoglobin S (Hb S) levels immediately prior to each transfusion during the measurement year (January 1 to December 31) (see the related "Numerator Inclusions/Exclusions" field).

Evidence Supporting the Measure

Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

Additional Information Supporting Need for the Measure

Sickle Cell Disease Prevalence and Incidence

Sickle cell disease (SCD) is one of the most common genetic disorders in the United States (U.S.) (Kavanagh et al., 2011). The National Heart, Lung and Blood Institute (NHLBI) (2002) estimates that 2,000 infants are born with SCD in the U.S. each year. SCD affects 70,000 to 100,000 children and adults in the U.S., predominantly those of African and Hispanic descent (Hassell, 2010).

Sickle Cell Disease Pathology and Severity

Vaso-occlusion (the sudden blockage of a blood vessel caused by the sickle shape of abnormal blood cells) is responsible for most complications of SCD, including pain episodes, sepsis, stroke, acute chest syndrome, priapism, leg ulcers, osteonecrosis and renal insufficiency (Steinberg, 1999). In addition, SCD can have hemolytic and infectious complications that result in morbidity and mortality in children with the condition (Kavanagh et al., 2011).

Sickle Cell Disease Burden in Daily Life

The effect of SCD on children and families is significant; severe pain episodes and hospitalizations restrict daily activities and reflect negatively on school attendance and performance, as well as on sleep and social activities (Lemanek, Ranalli, & Lukens, 2009; Alvim et al., 2005). Although medical management of SCD continues to improve over time, 196 children in the United States died from SCD-related causes between 1999 and 2002 (Yanni et al., 2009).

Sickle Cell Disease Cost

In a study of health care utilization among low income children with SCD between 2004 and 2007, 27% of these children required inpatient hospitalization and 39% used emergency care during a year. Of these children, 63% averaged one well-child visit per year and 10% had at least one outpatient visit with a specialist (Raphael et al., 2009). Patients with SCD use many parts of the health care system, incurring significant costs. In 2009, mean hospital charges for children with SCD and a hospital stay were \$23,000 for children with private insurance and \$18,200 for children enrolled in Medicaid (HCUPnet, Healthcare Cost and Utilization Project, 2012). Kauf et al. (2009) estimate the lifetime cost of health care per patient with SCD to be approximately \$460,000. Estimates place the cost of chronic transfusion therapy at up to \$400,000 per patient decade (Wayne, Schoenike, & Pegelow, 2000). Researchers have suggested that this expense, along with the associated risks of ongoing transfusion and its lack of permanent benefit, should be considered as part of any cost/efficacy analyses regarding alternative SCD therapies, including as hydroxyurea and bone marrow transplants (Wayne, Schoenike, & Pegelow, 2000).

See the original measure documentation for additional evidence supporting the measure.

Evidence for Additional Information Supporting Need for the Measure

Alvim RC, Viana MB, Pires MA, Franklin HM, Paula MJ, Brito AC, Oliveira TF, Rezende PV. Inefficacy of piracetam in the prevention of painful crises in children and adolescents with sickle cell disease. *Acta Haematol.* 2005;113(4):228-33. [PubMed](#)

Hassell KL. Population estimates of sickle cell disease in the U.S. *Am J Prev Med.* 2010 Apr;38(4 Suppl):S512-21. [PubMed](#)

HCUPnet. Healthcare Cost and Utilization Project. [Web site]. Rockville (MD): Agency for Healthcare Research and Quality; 2006-2009

Kauf TL, Coates TD, Huazhi L, Mody-Patel N, Hartzema AG. The cost of health care for children and adults with sickle cell disease. *Am J Hematol.* 2009 Jun;84(6):323-7. [PubMed](#)

Kavanagh PL, Sprinz PG, Vinci SR, Bauchner H, Wang CJ. Management of children with sickle cell disease: a comprehensive review of the literature. *Pediatrics.* 2011 Dec;128(6):e1552-74.

Lemanek KL, Ranalli M, Lukens C. A randomized controlled trial of massage therapy in children with sickle cell disease. *J Pediatr Psychol*. 2009 Nov-Dec;34(10):1091-6.

National Heart, Lung and Blood Institute (NHLBI). The management of sickle cell disease. 4th ed. Bethesda (MD): National Institutes of Health, National Heart, Lung and Blood Institute, Division of Blood Diseases and Resources; 2002 Jun. 188 p.

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). Basic measure information: hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease. Ann Arbor (MI): Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium; 2014 Apr. 39 p.

Raphael JL, Dietrich CL, Whitmire D, Mahoney DH, Mueller BU, Giardino AP. Healthcare utilization and expenditures for low income children with sickle cell disease. *Pediatr Blood Cancer*. 2009 Feb;52(2):263-7. [PubMed](#)

Steinberg MH. Management of sickle cell disease. *N Engl J Med*. 1999 Apr 1;340(13):1021-30. [PubMed](#)

Wayne AS, Schoenike SE, Pegelow CH. Financial analysis of chronic transfusion for stroke prevention in sickle cell disease. *Blood*. 2000 Oct 1;96(7):2369-72.

Yanni E, Grosse SD, Yang Q, Olney RS. Trends in pediatric sickle cell disease-related mortality in the United States, 1983-2002. *J Pediatr*. 2009 Apr;154(4):541-5. [PubMed](#)

Extent of Measure Testing

Reliability

Data and Methods. The testing data consisted of an audit of medical records from the three largest centers serving sickle cell disease (SCD) patients in Michigan during 2012: Children's Hospital of Michigan (CHM, Detroit), Hurley Medical Center (Hurley, Flint), and the University of Michigan Health System (UMHS, Ann Arbor). Combined, these sites treat the majority of children with SCD in Michigan. Medical records for all children with SCD meeting the measure specification criteria during the measurement year were abstracted at each site. Abstracting was conducted in two phases; during Phase 1, 435 records were abstracted among the three sites. In Phase 2, an additional 237 cases were abstracted at one site. In total, 672 unique records were reviewed for children with SCD to test this measure.

Reliability of medical record data was determined through re-abstractation of patient record data to calculate the inter-rater reliability (IRR) between abstractors. Broadly, IRR is the extent to which the abstracted information is collected in a consistent manner (Keyton et al., 2004). Low IRR may be a sign of poorly executed abstraction procedures, such as ambiguous wording in the data collection tool, inadequate abstractor training, or abstractor fatigue. For this project, the medical record data collected by two nurse abstractors were compared.

Measuring IRR at the beginning of the abstraction is imperative to identify any misinterpretations early on. It is also important to assess IRR throughout the abstraction process to ensure that the collected data maintain high reliability standards. Therefore, the IRR was evaluated during Phase 1 at each site to address any reliability issues before beginning data abstraction at the next site.

IRR was determined by calculating both percent agreement and Kappa statistics. While abstraction was still being conducted at each site, IRR assessments were conducted for 5% of the total set of unique patient records that were abstracted during Phase 1 of data collection. Two abstractors reviewed the same medical records; findings from these abstractions were then compared, and a list of discrepancies was created.

Three separate IRR meetings were conducted, all of which included a review of multiple SCD measures

that were being evaluated. Because of eligibility criteria, not all patients were eligible for all measures. Therefore, records for IRR were not chosen completely at random; rather, records were selected to maximize the number of measures assessed for IRR at each site.

Results. For this measure, 8 of 435 unique patient records from Phase 1 of the abstraction process were assessed for IRR across the three testing sites. Overall, these 8 records included 45 transfusions during the measurement year.

Table 5 of the original measure documentation shows the percent agreement and Kappa statistics for the measure numerator for each site and across all sites. The overall agreement for the measure is 98% and the Kappa is 0.95, indicating that a high IRR level was achieved.

Validity

Face Validity. The face validity of this measure was established by a national panel of experts and advocates for families of children with SCD convened by the Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). The Q-METRIC expert panel included nationally recognized experts in SCD, representing hematology, pediatrics, and SCD family advocacy. In addition, measure validity was considered by experts in state Medicaid program operations, health plan quality measurement, health informatics, and health care quality measurement. In total, the Q-METRIC SCD panel included 14 experts, providing a comprehensive perspective on SCD management and the measurement of quality metrics for states and health plans.

The Q-METRIC expert panel concluded that this measure has a high degree of face validity through a detailed review of concepts and metrics considered to be essential to effective SCD management and treatment. Concepts and draft measures were rated by this group for their relative importance. This measure was highly rated, receiving an average score of 8.2 (with 9 as the highest possible score).

Validity of Abstracted Data. This measure was tested using medical record data, which is considered the gold standard for clinical information; our findings indicate that these data have a high degree of face validity and reliability. We tested this measure among a total of 38 children younger than 18 years of age with sickle cell anemia (Table 6 of the original measure documentation). Overall, 45% of children with sickle cell anemia and on a chronic transfusion program received appropriate monitoring of Hb S levels immediately prior to transfusion (range: 0% to 52%).

Evidence for Extent of Measure Testing

Keyton J, King T, Mabachi N, Manning J, Leonard L, Schill D. Content analysis procedure book. Lawrence (KS): University of Kansas; 2004.

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). Basic measure information: hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease. Ann Arbor (MI): Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium; 2014 Apr. 39 p.

State of Use of the Measure

State of Use

Current routine use

Current Use

not defined yet

Application of the Measure in its Current Use

Measurement Setting

Ambulatory/Office-based Care

Hospital Outpatient

Professionals Involved in Delivery of Health Services

not defined yet

Least Aggregated Level of Services Delivery Addressed

Single Health Care Delivery or Public Health Organizations

Statement of Acceptable Minimum Sample Size

Unspecified

Target Population Age

Age less than 18 years

Target Population Gender

Either male or female

National Strategy for Quality Improvement in Health Care

National Quality Strategy Aim

Better Care

National Quality Strategy Priority

Prevention and Treatment of Leading Causes of Mortality

Institute of Medicine (IOM) National Health Care Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Data Collection for the Measure

Case Finding Period

The measurement year

Denominator Sampling Frame

Patients associated with provider

Denominator (Index) Event or Characteristic

Clinical Condition

Patient/Individual (Consumer) Characteristic

Therapeutic Intervention

Denominator Time Window

not defined yet

Denominator Inclusions/Exclusions

Inclusions

The eligible population for the denominator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program during the measurement year (January 1 to December 31).

Note:

Eligible children are restricted to those with sickle cell anemia, as determined by the hemoglobin variants identified in Table 1 of the original measure documentation, with the appropriate International Classification of Diseases, Ninth Revision (ICD-9) codes documented in the medical record.

Intake Period: January 1 of the measurement year through February 15 of the year following the measurement year.

Chronic Transfusion Program: Intervals between transfusions must be six weeks or less for the entire transfusion measurement year. The procedure codes for blood transfusions are identified in Table 2 of the original measure documentation.

Exclusions

Children with a diagnosis in the sampled medical record indicating one of the sickle cell disease (SCD) variants listed in Table 3 of the original measure documentation should not be included the eligible population *unless* there is also a diagnosis for a sickle cell variant listed in Table 1.

Exclusions/Exceptions

not defined yet

Numerator Inclusions/Exclusions

Inclusions

The eligible population for the numerator is the number of children younger than 18 years of age identified as having sickle cell anemia and on a chronic transfusion program who received monitoring of hemoglobin S (Hb S) levels immediately prior to each transfusion during the measurement year (January 1 to December 31).

Note:

Documentation in medical record must include, at a minimum, a note containing the date on which each treatment was administered.
Monitoring: Children receive a pre-transfusion Hb S level (blood draw) 24 hours or less prior to each transfusion (see Table 2 of the original measure documentation).

Exclusions

Unspecified

Numerator Search Strategy

Fixed time period or point in time

Data Source

Electronic health/medical record

Paper medical record

Type of Health State

Does not apply to this measure

Instruments Used and/or Associated with the Measure

Unspecified

Computation of the Measure

Measure Specifies Disaggregation

Does not apply to this measure

Scoring

Rate/Proportion

Interpretation of Score

Desired value is a higher score

Allowance for Patient or Population Factors

not defined yet

Standard of Comparison

not defined yet

Identifying Information

Original Title

Hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease.

Measure Collection Name

Sickle Cell Disease Measures

Submitter

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC) - Academic Affiliated Research Institute

Developer

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC) - Academic Affiliated Research Institute

Funding Source(s)

This work was funded by the Agency for Healthcare Research and Quality (AHRQ) and the Centers for Medicare & Medicaid Services (CMS) under the CHIPRA Pediatric Quality Measures Program Centers of Excellence grant number U18 HS020516.

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Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

Adaptation

This measure was not adapted from another source.

Date of Most Current Version in NQMC

2014 Apr

Measure Maintenance

Unspecified

Date of Next Anticipated Revision

Unspecified

Measure Status

This is the current release of the measure.

The measure developer reaffirmed the currency of this measure in January 2016.

Measure Availability

Source available from the [Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium \(Q-METRIC\) Web site](#) . Support documents are also available.

For more information, contact Q-METRIC at 300 North Ingalls Street, Room 6C08, SPC 5456, Ann Arbor, MI 48109-5456; Phone: 734-232-0657; Fax: 734-764-2599.

NQMC Status

This NQMC summary was completed by ECRI Institute on January 23, 2015. This NQMC summary was verified by the measure developer on March 2, 2015.

The information was reaffirmed by the measure developer on January 7, 2016.

Copyright Statement

This NQMC summary is based on the original measure, which is subject to the measure developer's copyright restrictions.

Inform Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC) if users implement the measures in their health care settings.

Production

Source(s)

Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium (Q-METRIC). Basic measure information: hemoglobin S monitoring prior to chronic transfusion among children with sickle cell disease. Ann Arbor (MI): Quality Measurement, Evaluation, Testing, Review, and Implementation Consortium; 2014 Apr. 39 p.

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